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Neural Substrates of Spelling Mechanisms: Evidence from Primary Progressive Aphasia

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Primary Progressive Aphasia (PPA) is a neurodegenerative disease that affects primarily language abilities. Spelling is amongst the first language abilities affected in PPA with prognostic and diagnostic value for the variant of PPA (Sepelyak et al., 2011). Recent studies have led to the following hypotheses regarding the neural networks that support two of the main spelling mechanisms: (1) the mechanism of access to the orthographic lexical representations for the spelling of known words depends in left inferior frontal gyrus and midfusiform gyrus (2) the mechanism of phoneme-grapheme conversion (PGC) for the spelling of unfamiliar words depends on left inferior frontal gyrus and supramarginal gyrus. Evidence for these neural networks is based in acute stroke studies (Philipose et al., 2007), case-studies of tumor resections (Tsapkini and Rapp, 2010), and a meta-analysis of chronic stroke studies and fMRI studies of healthy controls (Purcell et al., 2011). In the present study we aimed to evaluate these hypotheses in PPA in order to subsequently implement targeted neuromodulatory interventions for spelling in PPA.

We studied the full range of spelling mechanisms in 58 individuals with PPA using an extensive spelling battery (Johns Hopkins Dysgraphia Battery). Multiple regressions of cortical atrophy areas on performance in spelling tasks revealed that, atrophy in specific neural areas predicted performance in tasks reflecting distinct spelling mechanisms. These neural areas are: left inferior frontal gyrus for both the direct lexical access and the phoneme-grapheme conversion (PGC) mechanisms, left supramarginal gyrus for the PGC mechanism; and left fusiform gyrus for the direct access to the orthographic lexicon mechanism. Results thus confirmed our hypotheses. It seems we can now opt for neurally-targeted neuromodulatory interventions for spelling deficits.

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